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An olfactory neuroblastoma presenting as posterior reversible leukoencephalopathy syndrome

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ABSTRACT

Olfactory neuroblastomas are rare malignancies of the nasal and sinus cavities. They have been associated with paraneoplastic syndromes due to secretion of adrenocorticotropin hormone (ACTH) or antidiuretic hormone. These associated paraneoplastic syndromes can present with a wide variety of symptoms that can make diagnosis of the underlying tumor difficult. Here we present the case of a 23-year-old woman who had a seizure due to the development of posterior reversible leukoencephalopathy syndrome because of secondary hypertension due to Cushing's syndrome, which was in turn found to be due to ectopic ACTH production from a metastatic olfactory neuroblastoma.

KEYWORDS Adrenocorticotropin hormone; Cushing's syndrome; esthesioneuroblastoma; olfactory neuroblastoma; posterior reversible leukoencephalopathy

Olfactory neuroblastomas are rare, neural crest-derived malignancies that typically arise from olfactory epithelium around the cribriform plate.[1,2] They typically present with unilateral nasal congestion or obstruction, headache, sinusitis, and epistaxis, but diagnosis often takes months. Rarely, paraneoplastic syndromes can result from tumor secretion of hormones. Here, we report the case of Cushing's syndrome secondary to an adrenocorticotropin hormone (ACTH)-secreting metastatic olfactory neuroblastoma, complicated by posterior reversible leukoencephalopathy syndrome (PRES).

CASE REPORT

A 23-year-old woman was brought to the emergency department after having two seizures. She had been experiencing double vision for several days, had gained 30 pounds, and had a nasal polyp. Head computed tomography showed a left maxillary sinus tumor with extraocular muscle displacement. Subsequent magnetic resonance imaging (MRI) showed bilateral medial hemispheric signal abnormalities with focal areas of hemorrhage compatible with hemorrhagic PRES. The MRI also detailed the 6.3-cm left sinonasal mass with components in the left ethmoid and maxillary sinuses, nasal cavity, and nasopharynx, abutting but not displacing the optic nerve. The inferior and medial rectus muscles were both displaced, and the structure was abutting the left inferior frontal lobe with dural thickening along the left anterior cranial fossa without evidence of invasion. Additionally, a suspicious 1.3-cm enhancing lymph node was noted at the left angle of the mandible.

The patient had a Cushingoid appearance, with truncal obesity, extremity wasting, and abdominal striae. Random and morning cortisol levels were 99.3 mcg/dL and 101.5 mcg/dL, respectively, and 24-hour urine cortisol was above assay limit. Serum ACTH was 1001 pg/mL, inferring ectopic ACTH production, which was confirmed using a dexamethasone suppression test. Her cortisol levels improved with administration of ketoconazole. Spironolactone and lisinopril controlled her blood pressure, but she regularly required additional potassium supplementation to remain normokalemic. With the aforementioned MRI negative for structural pituitary pathology, tumor and lymph node biopsies were consistent with olfactory neuroblastoma, confirming Kadish Stage D metastatic olfactory neuroblastoma. Tumor samples stained strongly positive for ACTH, confirming clinical suspicions that this was an ACTH-secreting olfactory neuroblastoma.

The patient was given a 3-day course of induction chemotherapy with cisplatin and etoposide in an attempt to preserve

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her eye. This was done in accordance with previously published recommendations for chemotherapy administration in aggressive and locoregionally advanced disease.[3] She tolerated the regimen well and was discharged from the hospital with neutropenia but otherwise stable. Unfortunately, 2 days after discharge, the patient suffered a small laceration while shaving her left armpit and developed a superficial infection that rapidly progressed to necrotizing fasciitis across and down the left torso. Despite multiple emergent surgical interventions and intensive care, she died due to septic shock and *Serratia* bacteremia.

DISCUSSION

This is only the second documented case of a Kadish Stage D, metastatic ACTH-secreting olfactory neuroblastoma. While these tumors account for about 3% of all nasal cavity malignancies, most do not induce paraneoplastic syndromes. Management usually involves surgery and radiation, but patients with advanced disease or a contraindication to surgery have received chemotherapy (usually a cisplatin-based regimen or a combination cyclophosphamide, vincristine, and doxorubicin).[2] In this patient, it was deemed most appropriate to give neoadjuvant chemotherapy to try preserving vision in her left eye.

Cushing's disease/syndrome has been associated with PRES in only three case reports and only once in an adult.[4] This clinicoradiographic syndrome is characterized by headache, impaired consciousness, seizure, visual disturbances, or focal neurologic deficits with bilateral subcortical vasogenic edema affecting the parieto-occipital regions on imaging.[5] PRES is usually associated with a discernible trigger, especially blood pressure fluctuations, renal failure, eclampsia, exposure to immunosuppressive/cytotoxic agents, or autoimmunity.[5]

While this report highlights the 21st case of an ACTH-secreting olfactory neuroblastoma and the fourth case of Cushing's-related PRES, it is the first known occurrence of the three conditions interconnected in this manner.

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